

were continued. (Uthman BM et al. Outcome for West Syndrome following surgical treatment. Epilepsia Sept/Oct 1991; 32:668-671).

COMMENT. This report indicates that infantile spasms and hypsarrhythmia of a diffuse pattern may be associated with a focal cerebral lesion amenable to surgery. Infantile spasms associated with COFS syndrome (cerebro-oculo-facial-skeletal syndrome) in a 3-month-old child is reported from the Department of Neurology, New York Medical College, New York, NY (Harden CL et al. Pediatr Neurol July/Aug 1991; 7:302-304). COFS syndrome is a rare autosomal recessive condition characterized by microcephaly, microphthalmia and/or cataracts, neurogenic arthrogryposis and multiple congenital anomalies. The infantile spasms and hypsarrhythmia resolved during ACTH therapy.

DIAGNOSIS OF JUVENILE MYOCLONIC EPILEPSY

Factors contributing to the misdiagnosis of juvenile myoclonic epilepsy (JME) in an epilepsy clinic have been examined in 70 patients at the Division of Neurology, King Khalid University Hospital, Riyadh, Saudi Arabia. More than 90% were undiagnosed on referral and 33% were not recognized initially in the epilepsy clinic. The delay in diagnosis was 8 years from onset and 17 months from the first evaluation in the clinic. Factors responsible for the delayed diagnosis include the following: myoclonic jerks rarely reported by patients; generalized tonic-clonic seizures may be nocturnal without circadian relation to awakening from sleep; unilateral jerks may suggest simple partial seizures; absence seizures may antedate jerks and GTCS seizures by 4.5 years and are frequently unrecognized. The EEG was significant in confirming the diagnosis in 63% of patients. Valproate is considered the treatment of choice and clonazepam is used as an adjunctive treatment. (Panayiotopoulos CP et al. Juvenile myoclonic epilepsy: factors of error involved in the diagnosis and treatment. Epilepsia Sept/Oct 1991; 32:672-676).

COMMENT. Failure to recognize JME may result in improper choice of anticonvulsant therapy, resultant status epilepticus, and failure to provide appropriate genetic counseling. This study reemphasizes the atypical history in some cases and the frequency of occurrence of absence seizures as the initial manifestation.

MEMORY AND LEARNING DISABILITIES

ANATOMY OF MEMORY

Studies of the anatomy and function of the brain system for memory in humans and animal models are reviewed from the Veterans Affairs Medical Center, San Diego and the Department of Psychiatry, University of California, San Diego, La Jolla, CA. Patients who underwent temporal lobe surgery developed memory impairment only when the removal extended far enough posteriorly to include the hippocampus and the parahippocampal gyrus. Horel

proposed that memory functions were disrupted not by hippocampal damage, but by damage to temporal stem white matter adjacent to the hippocampus. Damage to both the hippocampus and the amygdala was required to produce severe amnesia in monkeys and humans. A circumscribed bilateral lesion involving the entire rostro-caudal extent of the CA1 field of the hippocampus caused memory impairment in the absence of other cognitive dysfunction. By use of the high resolution MRI the hippocampal region was found to be shrunken and atrophic in 4 amnesic patients while the temporal lobe was of normal size. Memory is classified as declarative (explicit) or non-declarative (implicit). **Declarative memory** refers to recollection of facts and events and depends on the integrity of the medial temporal lobe. **Non-declarative memory** refers to a collection of skills, habits, priming, conditioning and non-associated learning, all of which are non-conscious recollections, and is independent of the medial temporal lobe. The medial temporal lobe memory system is essential for establishing long-term memory for facts and events and is needed to bind together the distributed storage sites in neocortex that represent a whole memory. As time passes after learning, memory stored in the neocortex gradually becomes independent of the medial temporal lobe structures and the role of this system in memory is only temporary. More permanent memory develops presumably as a result of slow synaptic change and in concert with normal forgetting. The process by which the burden of long-term permanent memory storage is gradually assumed by the neocortex assures that the medial temporal lobe system is always available for the acquisition of new information. (Squire LR, Zola-Morgan S. The medial temporal lobe memory system. Science Sept 20, 1991; 253:1380-1385).

COMMENT: The article refers to a neuropsychological evaluation of a patient, H.M., with a profound and a selective impairment in human memory after bilateral surgical removal of the medial temporal lobe (Scoville WB, Milner B. J Neurol Neurosurg Psychiatry 1957; 20:11). A reference to Marcel Proust's novel of the mind "The Remembrance of Things Past," is made in an editorial by Hilts PJ in The New York Times Sept 24, 1991. Proust's memory of a town and gardens was triggered by the pleasures of a morsel of cake soaked in a spoonful of warm tea, a mechanism involving the amygdala.

EEG AND SPELLING DISABILITIES

The EEGs in 23 13-year-old Finnish-speaking boys with spelling disabilities and in 21 matched controls were studied in the Departments of Child Neurology, Paediatrics, Clinical Neurophysiology and Psychology, University of Helsinki, Finland. The visual assessment of the records showed an abnormal EEG in 48% of the index group and in 25% of controls. The abnormalities included a general excessive slow activity, temporal slow wave activity and non-paroxysmal slowing. The frequency of paroxysmal activity did not differ significantly in the two groups; 4% in the index group and 9.5% in controls. A neurotic disposition measured by the Tennessee Self-concept scale and clinical subscale was more frequent in the index group than controls and reading was slightly impaired. Arithmetic skills were comparable in the